MONOSTOTIC FIBROUS DYSPLASIA OF THE SPINE: CASE REPORT

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A case of monostotic fibrous dysplasia of the spine is presented. It was referred to us after a history of 3 years' pain. The site of the anomaly is so exceptional that it invited comparison with the six other cases described in the literature. This comparison produced resemblances which differentiate this particular type of monostotic fibrous dysplasia form that located at other sites, both in its clinical aspects and its radiographic appearances.

Fibrous dysplasia of bone is a congenital anomaly of development. It can appear in monostotic or polyostotic forms, and is characterised by abnormal development of both the fibrous and osseous constituents of bone. The term was first coined by Lichtenstein in 1938 and many case reviews of both the monostotic and polyostotic types have been reported in the literature (Lichtenstein and Jaffe, 1942; Schlumberger, 1946; Wells, 1949; Harris *et al.*, 1962; Stewart *et al.*, 1962; Reed, 1963; Firat and Stutzman, 1968; Henry, 1969; Campanacci and Leonessa, 1970; Gibson and Middlemiss, 1971).

In decreasing order of frequency, monostotic fibrous dysplasia affects the maxilla, proximal femur, tibia and humerus. The hand and foot are rarely affected. Vertebral localisation is rare (Campanacci, 1981); we found only 6 cases recorded in the literature. A case located in the third lumbar vertebra recently observed at the Tumour Centre of the Rizzoli Orthopaedic Institute is therefore considered worth recording.

CASE DESCRIPTION

The patient was a female aged 36 years who had complained of pain in the lumbar spine, particularly during flexion, for the previous 3 years. She had been treated medically but with only temporary relief. After 2 years the pain had increased following an appendicectomy. A radiographic examination at that time indicated structural alterations in the body of L_3.

Clinical examination revealed slight dorsal kyphosis and painless movement over about two thirds of the normal range. Hyperextension produced pain, as did the straight leg raising test at extreme ranges. Pain and paraesthesia occurred sporadically in the S_1 root distribution area.

A radiographic examination (Figs. 1a and b) showed osteolysis of the body of L_3 with a «ground glass» appearance, clearly demarcated posteri-

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orly, and slight swelling of the vertebral body anteriorly.

A CT scan clearly indicated (Figs. 2a and b) that the area of osteolysis interrupted the anterior cortex of the vertebral body, while posteriorly it was delimited by an obvious border of sclerotic bone.

Scintigraphy of the entire skeletal system (Tc 99 methylenediphosphonate) indicated hyperaccumulation exclusively confined to the 3rd lumbar vertebra (Fig. 2c). Further radiographic examination of the total skeletal system did not show any other structural alterations.

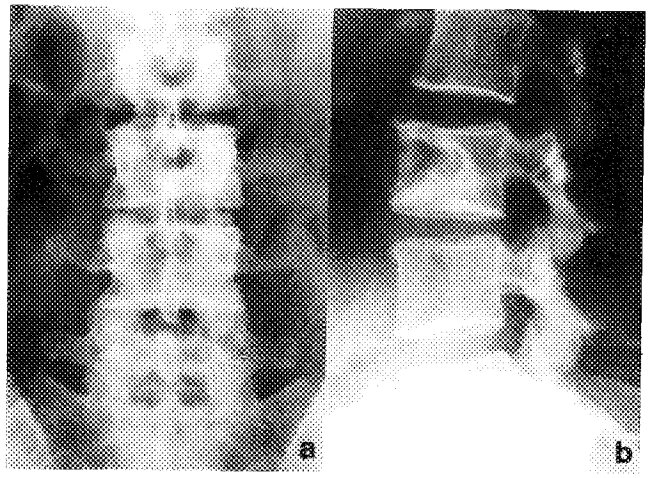

FIG. 1 - Radiograph of lumbar spine; monostotic fibrous dysplasia L_3. See description in text.

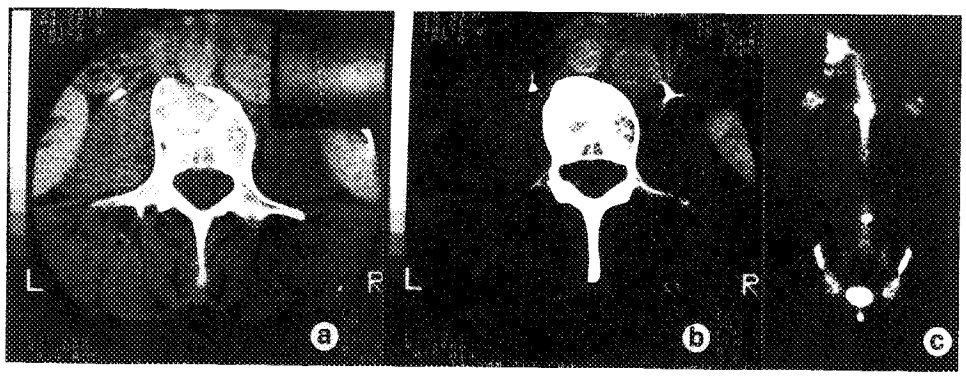

FIG. 2 - (a, b) CT scan at L_3 and (c) total body scintigraphy. See description in text.

An operation by the anterior approach was carried out. This consisted of curettage and the insertion of an autoplastic bone graft from the iliac crest. Surgery was indicated primarily in view of the need to ascertain the precise diagnosis by histological examination.

At operation a cavity was found in the body of L_3 containing greyish-white dense fibrous tissue resembling a scirrhus or compact desmoid tumour. Frozen section confirmed its benign nature, so curettage and grafting was completed.

Subsequent histological analysis confirmed the diagnosis of fibrous dysplasia. Detailed examination (Figs. 3a and b) showed diffuse proliferation of dense fibrous tissue in the medullary component with bone trabeculae surrounded by atypical fibrocytes. This fibrous component was arranged in layers and the cell nucleus was orientated along the major axis. The cellularity was intense and cytologically monomorphous.

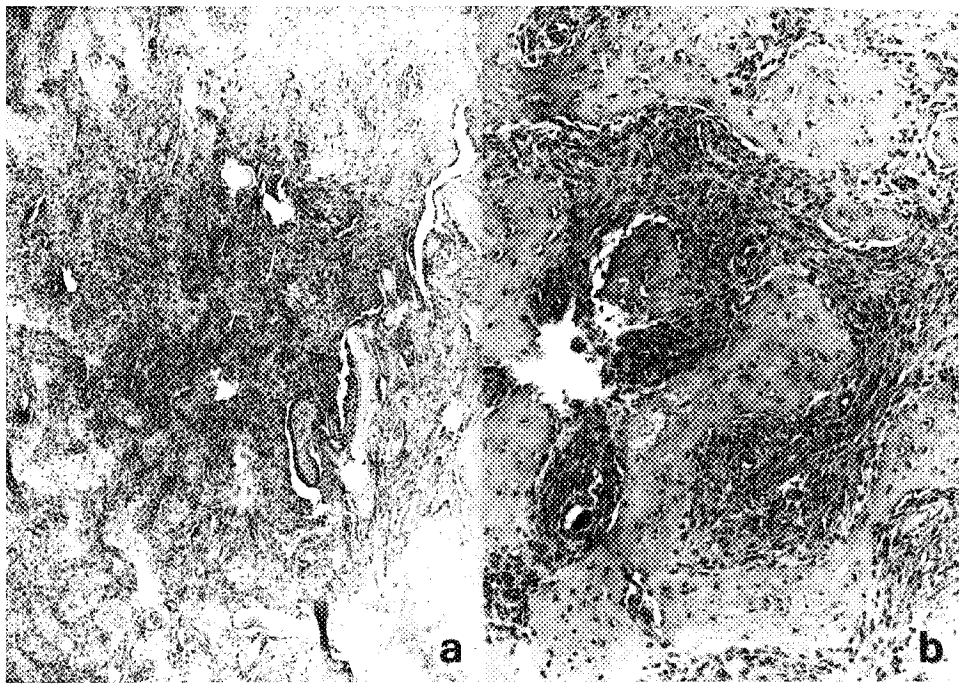

FIG. 3 - Histological examination of specimen. (a) 80x (b) 200x magnification. See description in text.

Fourteen months later the patient reported persistent pain in the lumbar spine. This had reappeared 5 months after operation and gradually became more insistent, involving radiation to the lower extremities without any precise metamerical distribution. A recent radiograph shows (Figs. 4a, b and c) good incorporation of the graft.

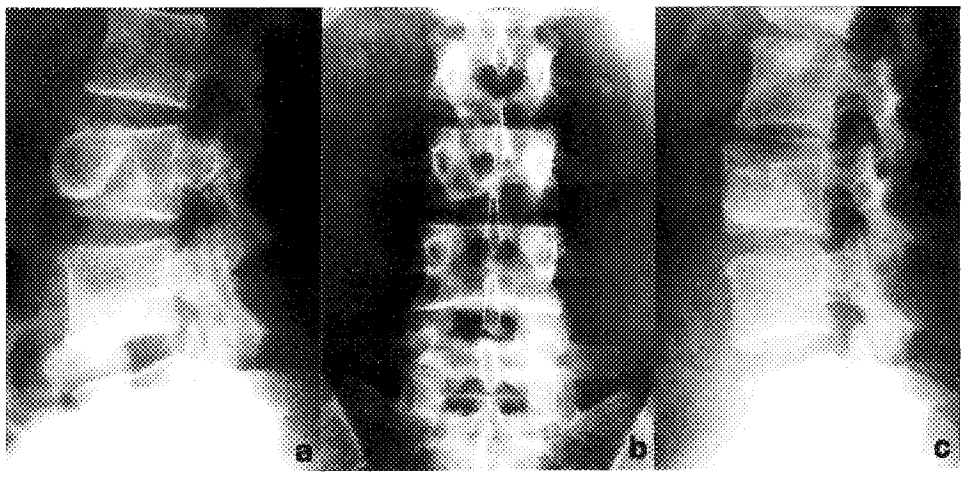

FIG. 4 - Postoperative radiograph: (a) showing autoplastic bone graft. (b, c) one year follow-up shows good incorporation and remodelling of vertebral body.

Table 1

Authors	Sex	Age	Site	Clinical features	Radiographic appearances	Treatment (follow-up)
Schlumberger, 1946	M	20	C4	Fracture. After one year radiograph positive.	Lysis of the body, peduncle and right lamina.	Biopsy
Ledoux-Lebard *et al.*, 1953	F	58	L1	For 6 years pain and medullary compression	Lysis of the body and peduncles	Biopsy + decompression
Rosendhal-Jensen, 1956	F	35	L4	Trauma. Pain after one year	Lysis of the body, facets, transverse processes and spinous processes	Curettage (one year: subject well). X-ray: calcificatio in lesion
Harris *et al.*, 1962	M	42	L4	Constant pain	Radiopaque lesion in transverse process	Biopsy (4 years: occasional pain)
Daniluk *et al.*, 1979	F	28	L4	Pain and dysuria for 10 years	Swelling of facets and transverse process	—
Resnik et al., 1984	F	27	C6	Acute onset of pain after trauma	Lysis of the body, peduncle and right lamina	Biopsy
Our case being examined	F	36	L3	Continuous pain for 3 years	Lysis of the body with anterior swelling	Curettage and graft. (At 14 months, residual pain but Rx shows good incorporation of graft)

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DISCUSSION

Monostotic fibrous dysplasia is uncommon but has a higher incidence than the polyostotic form. In our review of the literature we found one series where the ratio was inverted (Harris *et al.*, 1962). The cases observed at the Tumour Centre of the Rizzoli Orthopaedic Institute confirm the prevalence of the monostotic form, which was 67% of all cases of fibrous dysplasia in our records.

Localisation in the spine is exceptional. Of the 6 cases reported in the literature, 3 were in the lumbar region and 3 in the cervical spine. Dorsal and sacral localisations have not been described. Four cases were females and the ages ranged from 27 to 58 years. All cases had a long history of vertebral pain, sometimes with symptoms of medullary or radicular compression (Ledoux-Lebard and Soulquin, 1953; Harris *et al.*, 1962; Daniluk and Witwicki, 1979). At times the onset of symptoms was triggered by trauma, albeit slight (Schlumberger, 1946; Rosendhal-Jensen, 1956; Resnick and Lininger, 1984). The relevant data of these 6 cases, together with that of the present case, is summarized in Table 1.

Monostotic fibrous dysplasia is often asymptomatic and, as frequently occurs in the ribs, may only be discovered as a result of radiography carried out for other reasons (Wilner, 1982). In our review of the literature, the lesions were discovered after years of pain or after accident-related trauma before which the lesion had been symptomatically quiescent.

On radiographic examination the extent of vertebral involvement varied from localised osteolysis in a single transverse process (Harris *et al.*, 1962) to involvement of the body, facets, transverse and spinous processes (Rosendhal-Jensen, 1956).

In polyostotic fibrous dysplasia vertebral localisation occurs at the lumbar level in 14% of cases and at the cervical level in 7% of cases. In the monostotic form the problem of differential diagnosis may arise with regard to angioma or aneurysmal bone cyst, which may also appear radiographically as trabeculated swelling of the vertebral body. Osteoblastoma may also have to be considered since this also may be characterized by a diffuse radiopaque «frosted glass» appearance. Finally, fibrous dysplasia may be characterised by cartilaginous islands which are at times calcified. However, these have never been found in the vertebral localisation.

It is important to bear in mind the possible malignant transformation of both monostotic and polyostotic forms of the anomaly (1-2% of cases: Campanacci, 1981). Osteosarcoma is the most frequent, but fibrosarcoma and chondrosarcoma have also been recorded.

Surgical treatment is only indicated in the symptomatic forms of the anomaly, in particular, in the presence of neurological deficit, or for the prevention and correction of vertebral deformity due to pathological fracture and consequent axial deviation of the vertebral column.

Radiation therapy is contraindicated because of its ineffectiveness and the risk of sarcomatous transformation. For this reason, an exact differential diagnosis must be made with regard to angioma or aneurysmal bone cyst, both of which may be successfully treated by radiation therapy.

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